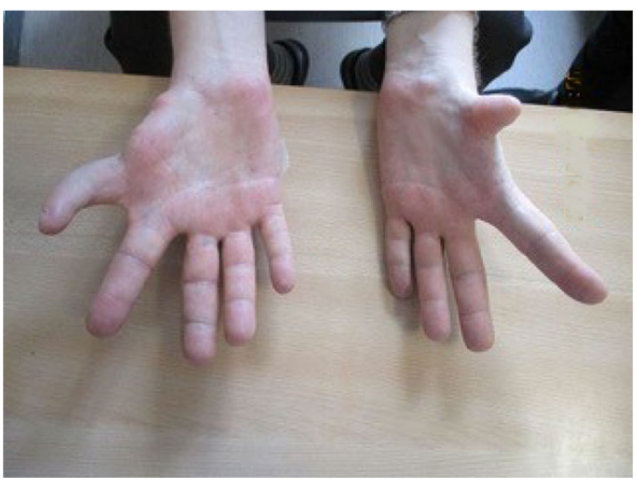
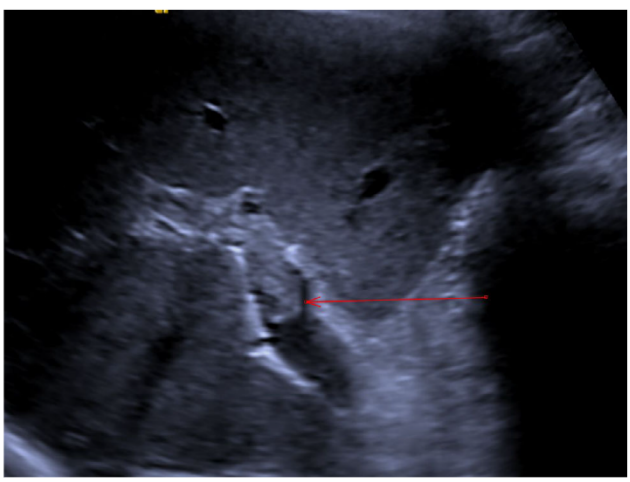
Management of a patient with a rare congenital limb malformation syndrome after SARS-CoV-2 vaccine-induced thrombosis and thrombocytopenia (VITT)





A 54-year-old man with a rare congenital limb malformation (left image) presented to the Accident and Emergency department with a 7-day history of worsening headache, bruising and unilateral right calf swelling. There was a strong family history of a rare congenital limb deformity, his mother and his maternal grandfather having the same congenital deformity. Computed tomography (CT) showed extensive cerebral venous sinus thrombosis and ultrasonography confirmed concurrent venous thrombosis in the portal vein (right image, filling defect indicated by arrow). There was also thrombophlebitis of the right leg. On presentation, his platelet count was 34 × 10^9/l and D-Dimer was 60 000 ng/ml. His blood film confirmed a true thrombocytopenia, with the absence of polychromasia, spherocytes and fragments making the diagnosis of thrombotic thrombocytopenic purpura unlikely. The patient had received the AstraZeneca SARS-CoV-2 vaccine 3 weeks before presentation, making the likely diagnosis vaccine-induced thrombosis and thrombocytopenia (VITT). An anti-platelet factor 4 (anti-PF4) antibody assay was 2·509 (normal range 0–0·4), thus confirming the diagnosis.

The patient was managed according to the current British Society of Haematology guidance with therapeutic intravenous immunoglobulin (IvIg) and anticoagulation,[1] although his limb malformation complicated management. The initial anticoagulant selected was danaparoid due to its immediate therapeutic effect and availability out of hours. Four days after receiving IvIg, his platelet count had increased to 204 × 10^9/l and D-dimer had fallen to 4400 ng/ml. On discharge, a direct oral anticoagulant (DOAC) was prescribed as the patient was unable to self-inject alternative agents owing to the congenital malformation of his hands. Warfarin was considered but deemed unsuitable as the patient could not travel for regular monitoring.

The evidence base for the long-term management of VITT is currently limited. As mechanistic understanding of these episodes improves, therapy can be tailored to the individual underlying circumstances.

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Reference

1. Guidance from the Expert Haematology Panel (EHP) on Covid-19 Vaccine-induced Immune Thrombocytopenia and Thrombosis (VITT). [Cited: 2021 May 28]. Available from: https://b-s-h.org.uk/media/19590/guidance-version-17-on-mngmt-of-vitt-20210420.pdf.

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